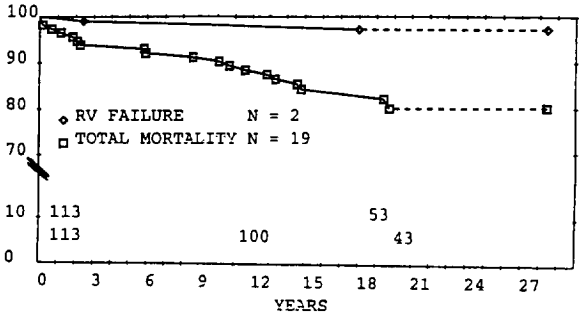
1982 were followed by serial cardiological review and a recent symptom questionnaire. Follow-up of late survivors was 10–28 years; 40 survive >20 years postoperatively. Right ventricular failure (RVF) was diagnosed if there were clinical signs and/or supportive radiological, echocardiographic or radionuclide evidence of progressive right heart dysfunction. Non-progressive RV dilatation was labelled RV dysfunction. *Results:* Actuarial survival was 93%, 90%, 84%, 80% and 80% at 5, 10, 15, 20 and 24 years respectively. Causes of the 19 deaths were: baffle obstruction 4; sudden, presumed arrhythmia 6; pulmonary vascular disease 2; miscellaneous 5; and RVF 2.



Although RV dysfunction is frequently observed, actuarial freedom from RVF is 97% at 18 years.

Conclusion: Progressive right ventricular failure is rare following the Mustard operation for simple transposition with intact septum.

10:45

702-2 Right Ventricular Function in Adolescents and Young Adults After Previous Mustard Operation

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The long term status of right ventricular function after intra-atrial baffle repair (Mustard) for transposition of the great arteries (TGA) was evaluated. Right ventricular ejection fraction (RVEF) and tricuspid regurgitation (TR) were estimated by radionuclide angiocardiography in 58 pts who were at least 10 yrs post-Mustard (mean age, 14 ± 3 yrs). Mean RVEF of 0.53 ± 0.10 did not differ from our normal. However, in 9 pts the most recent RVEF was <0.42, placing them >2SD below normal. Serial RVEF in 32 pts fell from a group mean of 0.54 ± 0.11 to 0.51 ± 0.11 ($p < 0.1$) over 9 ± 2 yrs. In 10 of these 32, RVEF either dropped to <0.42 or decreased >0.10 between studies. Significant TR was present in 27/52, and increased in 8 studied serially. This data demonstrates 1) though group RVEF in adolescent and young adult pts with previous Mustard is normal, almost 17% of pts have depressed function; 2) RVEF often worsens when studied serially in older pts; and 3) TR is significant in over half the previously operated pts. Though most of these post-operative TGA pts are free of clinical symptoms through adolescence, they are at significant risk for deterioration of function.

11:00

702-3 Factors Associated With Arrhythmias After Mustard Operation—30 Year Experience with 534 Patients

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To determine factors associated with survival and arrhythmias, we reviewed 534 (359 males) consecutive patients after Mustard operation (MUST) for transposition of the great arteries (TGA) from 1963 to 1993. TGA was isolated for 354 pts, associated with VSD in 73, pulmonary obstruction (PS) in 50, VSD with PS in 46 and complex anomalies in 11. Median age at MUST was 16 months (range, 21 days to 19 yrs). There were 52 hospital deaths (9.7%). Late death occurred in 76 of 482 operative survivors (15.8%) with up to 31 years follow-up (mean 12.6 ± 6.8 yrs). Late survival was 89% at 5 yr, 88% at 10 yr, 82% at 15 yr and 76% at 20 yr. Mode of late death included: Sudden, 39%; myocardial failure, 27%; pulmonary vascular disease, 10%; reoperation, 7%; pulmonary venous obstruction, 4%; non-cardiac, 4%; and other, 9%. Cox's proportional hazard modelling was used to assess independent factors associated with late death. Significant factors included: earlier date of operation (Odds Ratio, OR, 0.75 per 5 yrs), VSD closure at MUST (OR 2.5), and peri-operative atrial tachyarrhythmias at MUST (OR 1.9). Instantaneous risk (hazard) of late death declined, with a late peak and decline. Loss of sinus rhythmn (LSR) was noted in 29% of survivors: freedom from LSR was 82% at 5 yr, 67% at 10 yr, 60% at 15 yr and 52% at 20 yr. Significant independent factors associated with LSR included: pre-MUST surgical atrial septectomy (OR 1.7), peri-operative bradyarrhythmias (OR 1.7) and occurrence of atrial flutter (AFL) during follow-up. Instantaneous hazard of LSR showed a gradual decline after MUST with a second late peak. AFL was noted in 15% of survivors; freedom from AFL was 92% at 5 yr, 89% at 10 yr, 82% at 15 yr and 71% at 20 yr. Patients who were on digoxin vs. no medication at the time of their first AFL episode were less likely to have symptoms of presyncope or syncope (7% vs. 33%, $p = 0.06$). Independent factors associated with late AFL included: LSR (OR 2.6) and peri-operative arrhythmias including AFL (OR 3.1), bradyarrhythmias (OR 2.5) and permanent heart block (OR 12.2). Instantaneous hazard of AFL showed a gradual incline with a late peak. *Conclusion:* Late peaks in the risk of death, LSR and AFL necessitate ongoing follow-up in this population.

11:15

702-4 The Fontan Procedure for Pulmonary Atresia and Intact Ventricular Septum (PA and IVS): Operative and Late Results

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From 1973 through January 1, 1994, 860 Fontan procedures had been performed at the Mayo Clinic with 39 (4.5%) having been done on patients with PA and IVS. Age at surgery ranged from 1–10/12 to 21 years (median = 6). Thirty-eight of the 39 patients had had a total of 71 prior palliative cardiovascular procedures. Tricuspid valve (TV) and right ventricular (RV) size ranged from "minuscule" to a maximum of 55% of expected. Only four patients had significant right ventricular to coronary artery fistulae and only one of these was "right ventricular coronary dependent".

There were three operative deaths (7.7%) and have been two sudden unexpected late deaths (2-½ and 8 years post Fontan). One patient had a successful cardiac transplant seven years post Fontan. Present ages of survivors ranged from 3 to 29 years (median = 11.5 years) with a median post Fontan follow-up of 5.3 years. At recent follow-up, 33 of 34 survivors were in good or excellent condition and one was fair.

Conclusion: We feel patients with PA and IVS are not candidates for a conventional two ventricle repair unless the TV and RV are at least 70% of expected size. In patients with dimensions between 50% and 70%, a two ventricle repair combined with a bidirectional cavopulmonary anastomosis may be feasible. In patients with TV and RV size less than 50% of expected, the Fontan procedure provides "definitive palliation" and early and late results of this approach are encouraging.

11:30

702-5 Improved Survival After Fontan Operation for 62 Heterotaxy Patients

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The modified Fontan operation (MF) is the preferred palliative procedure for heterotaxy pts (HP) with complex cyanotic heart disease. Early mortality (EM) after MF has been reported to be greater for HP. In our recent experience (1987 – 92) EM after MF has improved dramatically. In order to determine if these improved results applied to HP, we reviewed all HP who had a MF at our institution between 1973 – 92 (n = 111). From 1973–92, 111/839 (13%) pts having a MF were HP [54 (6.5%) polysplenia, 57 (6.8%) asplenia]. HP were significantly older than non-HP at the time of MF (median age = 11 yrs vs 9 yrs; $p < 0.05$). We compared EM after MF for HP in two time periods: early [1973–86 (n = 49)] and recent [1987–92 (n = 62)]. EM decreased significantly in the recent group [9/62 (14.5%)] compared to the early group [21/49 (43%); $p < 0.001$]. In our early experience, asplenia pts had increased EM compared to polysplenia pts (65% vs 23%) but, recent experience shows that EM is similar for these two subsets of HP [asplenia: 5/34 (15%) vs polysplenia: 4/28 (14%)]. Many factors, including use of an intra-atrial tunnel, have contributed to these improved results.

Despite the recent reduction in EM for HP, this group remains at higher risk after MF than pts with tricuspid atresia (EM = 4%) or double inlet left ventricle (EM = 4%). This is most likely due to increased frequency of complicating features present in HP compared to non-HP. For example, atrioventricular valve (AVV) repair/replacement, a recognized risk factor for EM, was more frequent in HP than non-HP [28/62 (45%) vs 40/277 (14%); $p < 0.001$]. In HP who did not have AVV procedures EM was 9% (3/34) compared to 21% (6/28) in HP who had AVV procedures. The 9% EM rate observed in HP without AVV procedures is not significantly different from the 4% EM rate observed in non-HP ($p = 0.2$). *Conclusion:* Survival after MF for HP has improved dramatically. Although HP still experience a greater overall operative risk, some subgroups had EM rates which were not significantly different from non-HP.